*Thorax* 1986;41:66–67

# Short reports

## Adenoidal tissue as an aid to the diagnosis of sarcoidosis in childhood

R WILSON, M SWEATMAN, IS MACKAY, DN MITCHELL

*From the Brompton Hospital, Cardiothoracic Institute, and Medical Research Council Tuberculosis and Chest Diseases Unit, London*

The diagnosis of sarcoidosis in children and adults rests on the presenting symptoms and signs, with other available evidence, including the demonstration of non-caseating epithelioid and giant cell granulomas in biopsy material and a positive Kveim test reaction. Recent studies have shown that sarcoidosis is more prevalent among children than was previously recognised.[1-3] In childhood biopsy may present special problems since clinical evidence pointing to extrathoracic disease may be absent and fibreoptic bronchoscopy impracticable. We report our findings in two children in whom removal of apparently normal adenoidal tissue provided support for a diagnosis of sarcoidosis and thereby facilitated management.

### Case reports

CASE 1

A 15 year old Portuguese girl presented with gritty red eyes, which settled spontaneously. For two months she had also suffered intermittent central and upper abdominal pain, constipation, and low back pain. There was a history of adenotonsillectomy at 6 years and probable primary pulmonary tuberculosis at 2 years.

Two months later her eyes again became red and painful. They improved with topical atropine and steroid eye drops, but quickly deteriorated again while she was still having treatment. Examination showed anterior and posterior uveitis. There was no other clinical abnormality.

Investigations showed: haemoglobin 11·1 g/dl, hypochromic microcytic indices, white blood cell count 6·8 × $10^9$/l with normal differential count, erythrocyte sedimentation rate (ESR) 38 mm in one hour (Westergren). The following investigations yielded negative or normal results: urea and electrolytes, liver function, serum calcium, 24 hour urinary calcium, serum immunoglobulins, Rose-Waaler test, serum angiotensin 1 converting enzyme (ACE), toxoplasma dye test, Wasserman reaction, viral antibody titres, agglutination test for *Yersinia*, radiographs of lumbar spine and sacroiliac joints, barium meal and follow through and barium enema, sigmoidoscopy and histological examination of rectal biopsy material, and tuberculin (100 IU PPD), candidin, and trychophytin skin tests. A chest radiograph showed only a calcified lesion in the right upper zone.

Address for reprint requests: Dr R Wilson, The Host Defence Unit, Cardiothoracic Institute, Brompton Hospital, London SW3 6HP.

*Accepted 12 July 1985*

Adenoidal tissue was still present, despite previous surgery, and a further resection was carried out. Histologically the superficial lymphoid tissue was normal, but the deeper lymphoid and connective tissue contained numerous epithelioid and giant cell granulomas.

Because of diminishing visual acuity treatment with prednisolone (60 mg daily) was started, with rapid improvement in ocular signs and symptoms; the haemoglobin concentration rose to 13 g/dl and the ESR fell to 9 mm in one hour. A Kveim test, performed after steroid treatment had been started, gave a clinically and histologically negative result at four weeks.

CASE 2

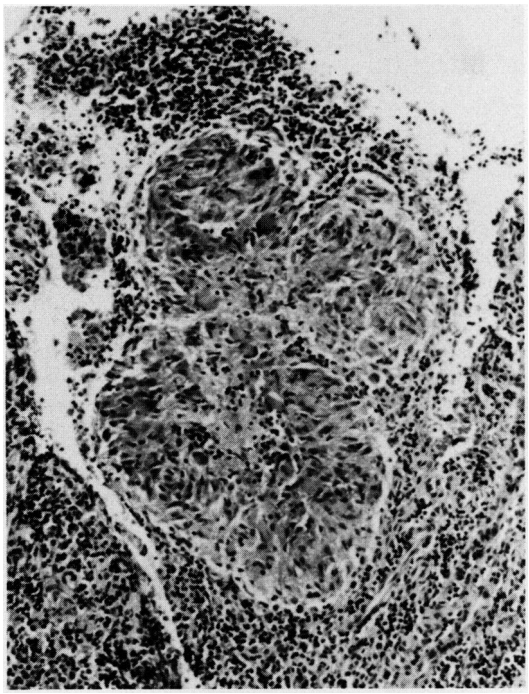
A 5 year old boy born in England of Nigerian parents presented with gritty red eyes. Bilateral lacrimal gland swelling had been present for two months. *Actinobacter* was cultured from an eye swab, and chloramphenicol eye drops produced rapid resolution of the redness and pain.

Over two months he deteriorated and developed transient intermittent fever, lethargy, and anorexia, with exertional dyspnoea and pleuritic left chest pain. On examination there was no abnormality apart from lacrimal gland enlargement.

Investigations showed: haemoglobin 9·1 g/dl, hypochromic microcytic indices, sickle cell trait positive, white cell count 11·5 × $10^9$/l (normal differential), ESR 150 mm in one hour (Westergren). Urea, electrolytes, liver function, serum calcium, 24 hour urinary calcium, latex rheumatoid factor, autoantibodies, and serum ACE were normal. A chest radiograph showed bilateral hilar lymphadenopathy with a diffuse reticulonodular infiltration throughout both lung fields. A Mantoux test (100 IU PPD) gave a negative result, and no acid fast bacilli were detected by microscopic examination or culture of urine and gastric aspirate on three occasions. No haemosiderin laden macrophages were found in a gastric aspirate.

Four weeks later the pulmonary infiltration was more noticeable and parotid swelling had developed. The nasopharynx was normal but in the absence of other available tissue an adenoidectomy was performed. Histological examination showed epithelioid and giant cell granulomas (fig). A Kveim test at four weeks was histologically positive.

His symptoms improved spontaneously without treatment. The lacrimal and parotid gland enlargement subsided within seven months, and the chest radiograph returned to normal within 13 months.



*Sarcoid granulomas in adenoidal tissue from case 2. (Haematoxylin and eosin, × 250.)*

## Discussion

We are unaware of any earlier reports of the use of adenoidal tissue to support a diagnosis of sarcoidosis; moreover, such an invasive procedure could be considered only when rapid confirmation is needed and no other site is accessible for biopsy. A Kveim test requires a minimum delay of four weeks after injection, and, as in case 1, the reaction is usually suppressed by steroid treatment.[4] Increased serum ACE activity is more frequent in children than in adults,[5] but in both our cases the level was normal.

Schaumann in 1914[6] described granulomas in tonsillar tissue in sarcoidosis, but subsequent studies of tonsillar biopsy material in this condition have been inconclusive.[7-9] On gross examination there is usually no abnormality, although Siltzbach and Blaugrund[9] reported two patients with large, pale tonsils that contained sarcoid granulomas.

Adenoidectomy has a lesser risk of haemorrhage than biopsy, can detect deep seated granulomas, and provides a useful option for histological confirmation of childhood sarcoidosis. Nasopharyngeal granulomas may be attributable to many causes, such as tuberculosis, leprosy, Wegener's granulomatosis, and cholesterol granulomas.[10] Other causes include congenital and acquired syphilis, scleroma, and fungal infection. Nasopharyngeal neoplasms may be associated with a granulomatous reaction in the draining lymphoid tissue. The differential diagnosis, however, presents little difficulty when the clinical picture is otherwise compatible with sarcoidosis.

We are very grateful to Professor O Wolff (Hospital for Sick Children, Great Ormond Street) and Dr M Silverman (Hammersmith Hospital) for referring these patients and for their kind permission in allowing us to report them.

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